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Misleading Leads: Focal Xanthogranulomatous Pyelonephritis in Childhood

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Xanthogranulomatous pyelonephritis is a morphologic variant of pyelonephritis. Focald-isease is very rare and can be misdiagnosed.

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CASE REPORT

A 6 1/2-year-old boy was admitted because of fever for 1 month before admission that did not resolve with antibiotics. The first ultrasound examination showed a large heterogeneous mass in the inferior pole of the right kidney with features suggesting a neoplasm.

At admission, the child had a large renal heterogeneous encapsulated mass without calcification that was considered at ultrasound examination and at CT to be a nephroblastoma (Fig. 1). There was a marked inflammatory syndrome with elevated white cells (20.900 per mm^3) and 79% polynuclear cells, with the C-reactive protein elevated to 73 mg/ml. There were, however, no signs of local or generalized infection specifically. There was no tenderness or pain in the right flank. Urine cultures also were negative and urine catecholamines levels were not elevated. Antibiotics (Amoxilline) and anticancer chemotherapy were initiated. Dactinomycin (4 daily doses of 15 mcg/kg) and vincristine (1.5 mg/m^2 per week) were started the day of the CT, with one cycle of dactinomycin and four weekly doses of vincristine completed within the first month. Steroids were not given. Very quickly after 8 days, the inflammatory syndrome disappeared. A follow-up CT scan (33 days after the first one) at the end of chemotherapy showed only a small residual wedge-shaped lesion (Fig. 2). The size of the tumor had decreased by 80%.

Right nephrectomy was performed 2 days after the second CT without intra-operative biopsy and the pathologist concluded that the patient suffered from focal xanthogranulomatous pyelonephritis (XPN) with perirenal inflammation (Figs. 3, 4). There was no underlying nephroblastoma.

DISCUSSION

This case points up two problems.

Difficulties in Diagnosis

It is not uncommon for patients suffering from nephroblastoma to have fever at the beginning of the disease. However, an associated inflammatory syndrome is rare. Retrospectively, there were also some features on the CT scan at admission that were not consistent with Wilms tumor. Most such tumors are more sharply defined and encapsulated than the lesion shown in Figure 1. The follow-up scan 33 days later showed a wedge-shaped aspect which again would be unusual for Wilms tumor, as the lesion seemed to follow the pattern of normal kidney anatomy. This is much more suggestive of an inflammatory process. Indeed, a clue to the correct diagnosis in cases like ours may be the ill-defined margins with inflammatory infiltration of the perinephric fat as is seen in Figure 1. Also, in focal inflammatory diseases, CT scans usually demonstrate tissue destruction in one pole often of low attenuation that fails to enhance after injection of contrast [2,4–6]. The rest of the kidney appears normal, but cases have been reported in which a clearly malignant renal tumor is accompanied by a pathologically proven pyelonephritis [1,3]. Percutaneous biopsy may be useful for diagnosis; it is done in France only if a benign tumor is suspected, because French centers generally follow the policy of the International Society of Pediatric Oncology. That policy is to start chemotherapy in suspected Wilms tumor patients without histologic confirmation of the diagnosis except in cases that demonstrate atypical clinical features.

Mechanisms to Explain the Decrease of the Focal Pyelonephritis

Wilms tumors often respond dramatically and very rapidly to chemotherapy, so that the response of the XPN

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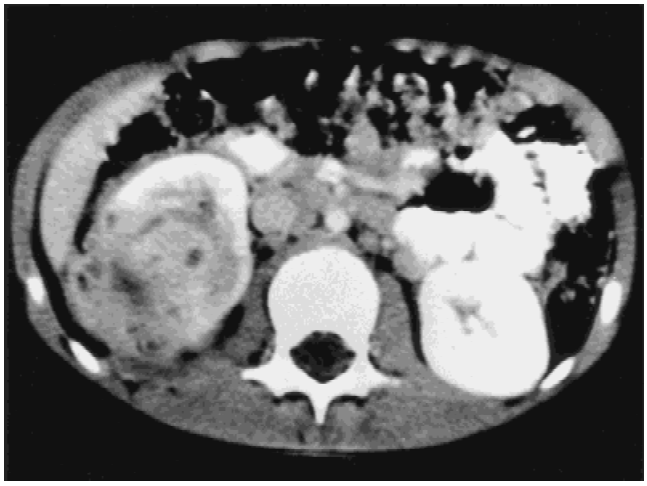

Fig. 1. CT cut: Large renal heterogeneous encapsulated mass of the right kidney without calcification.

Fig. 2. CT cut: 33 days later. Small wedge-shaped lesion of the right kidney.

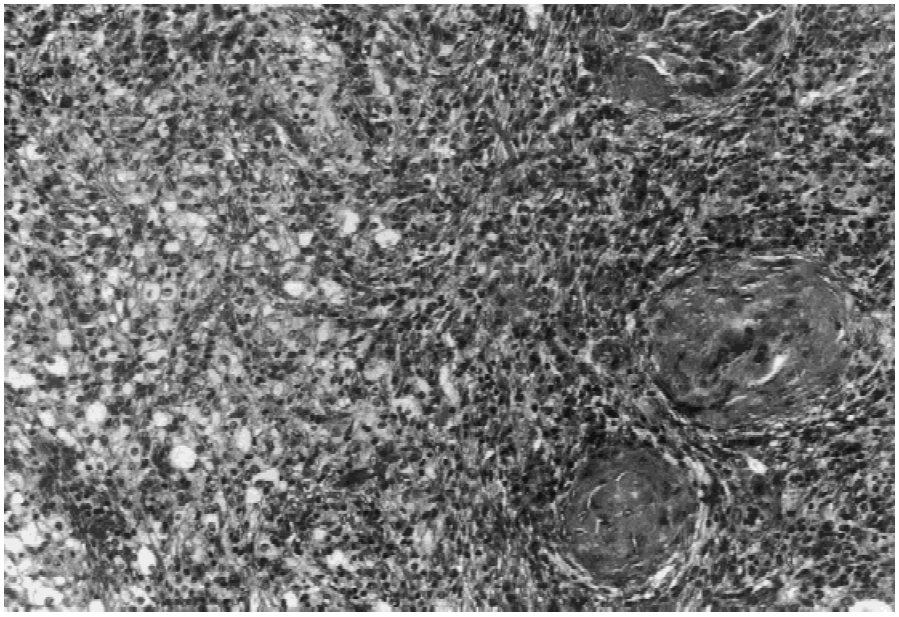

Fig. 3. Focal xanthogranulomatous pyelonephritis: replacement of renal tissue by inflammatory cells. Hematoxylin-eosin-saffron (HES), ×20.

in our patient was misleading. One possible explanation for the decrease in size of the infectious renal lesion is that a chronic XPN was in a stage of acute flare-up at the time of the initial examination and that the inflammation resolved spontaneously. It could also be related to the anti-cancer chemotherapy given, but one can only speculate as to the reason a pyelonephritic mass should have responded. The lesion is made up of lipid-laden macrophages with some lymphocytes. These cells are responsive to anti-mitotic agents. It is therefore a reasonable hypothesis that the treatments given for Wilms tumor caused cytolysis of noncancerous but proliferating and thus chemosensitive cells.

CONCLUSION

Masses that respond to anti-cancer drugs are not necessarily malignant in nature. Our experience with XPN presenting as a renal mass indicates that due attention must be paid to clinical signs and findings on imaging studies that suggest alternate diagnosis.

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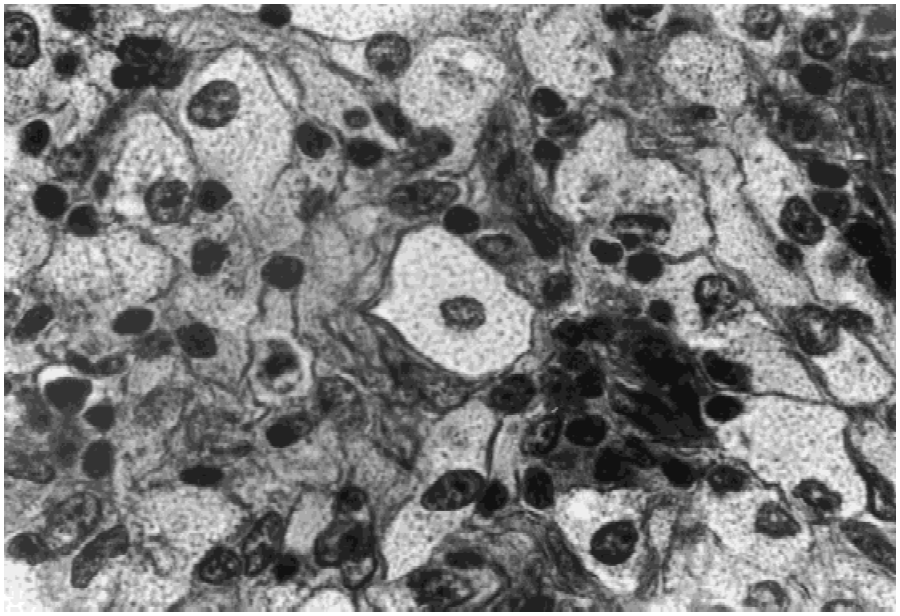

Fig. 4. Inflammatory cells are lipid-loaded macrophages admixed with some lymphocytes. HES, ×100.

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